Amendments to the Claims:

This listing of claims replaces all prior versions and listings of claims in the application:

Listing of Claims:

1. –34. (Cancelled)

35. (Amended) A kit for diagnosing a subject at risk for a hearing disorder, comprising: ~~one or more~~ at least two nucleic acid primers which hybridize under stringent conditions to a nucleic acid sequence ~~comprising~~ of SEQ ID NO: 1 or complement thereof, wherein the ~~primer~~ ~~amplifies~~ primers amplify all or a portion of exons 4 and 5 of SEQ ID NO:1 such that one or more nucleotides encoding one or more of an amino acid at residue 51, an amino acid at residue 66, an amino acid at residue 88 and an amino acid at residue 117 of SEQ ID NO:2 is amplified; and

and instructions for a diagnosing hearing disorder by detecting ~~a lesion which is an insertion, a deletion, or~~ a substitution of one or more nucleotides encoding one or more of a proline at residue 51 of SEQ ID NO:2, a valine at residue 66 of SEQ ID NO:2, a glycine at residue 88 of SEQ ID NO:2, and a tryptophan at residue 117 of SEQ ID NO:2.

36. The kit of claim 35, wherein the hearing disorder is DNFA9.

37. (Amended) The kit of claim 35, further comprising a nucleic acid probe which hybridizes under stringent conditions to the complement of SEQ ID NO:1, or ~~naturally occurring variants thereof which comprises~~ or a nucleic acid sequence that differs from SEQ ID NO:1 ~~lesion~~ at one or more nucleotides encoding one or more of a proline at residue 51 of SEQ ID NO:2, a valine at residue 66 of SEQ ID NO:2, a glycine at residue 88 of SEQ ID NO:2, and a tryptophan at residue 117 of SEQ ID NO:2~~, wherein~~

~~the probe hybridizes to a portion of the complement of SEQ ID NO:1, or the naturally occurring variant, such that a lesion at one or more nucleotides encoding a proline at residue 51 of SEQ ID NO:2, a valine at residue 66 of SEQ ID NO:2, a glycine at residue 88 of SEQ ID NO:2, or a tryptophan at residue 117 of SEQ ID NO:2 is detected~~ and which detects the absence or presence of a substitution at one or more nucleic acids that encode the proline at residue 51 of SEQ ID NO:2, the valine at residue 66 of SEQ ID NO:2, the glycine at residue 88 of SEQ ID NO:2, or the tryptophan at residue 117 of SEQ ID NO:2.

38. (Previously presented) The kit of claim 37, wherein the kit comprises more than one probe.

39. (Previously presented) The kit of claim 37, wherein the probe is a labeled probe.

40. (Previously presented) The kit of claim 38, wherein one or more of the probes is a labeled probe.

41. (Previously presented) The kit of claim 37, wherein the primer is at least 12 nucleotides in length.

42. (Previously presented) The kit of claim 37, wherein the primer comprises at least 12 consecutive nucleotides of SEQ ID NO:1.

43. (Amended) A kit for diagnosing a subject at risk for a hearing disorder, comprising: one or more nucleic acid probes which hybridize under stringent conditions to the complement of SEQ ID NO:1 or ~~naturally occurring variants thereof which comprises a lesion~~ a nucleic acid sequence that differs from the SEQ ID NO:1 ~~lesion~~ at

one or more nucleotides encoding one or more of a proline at residue 51 of SEQ ID NO:2, a valine at residue 66 of SEQ ID NO:2, a glycine at residue 88 of SEQ ID NO:2, and a tryptophan at residue 117 of SEQ ID NO:2~~, wherein the probe hybridizes to a portion of the complement of SEQ ID NO:1 or the naturally occurring variant such that a lesion at~~that includes ~~one or more nucleotides encoding: a proline at residue 51 of SEQ ID NO:2, a valine at residue 66 of SEQ ID NO:2, a glycine at residue 88 of SEQ ID NO:2, or a tryptophan at residue 117 of SEQ ID NO:2 is detected~~ and which detects the absence or presence of a substitution at one or more nucleic acids that encode the proline at residue 51 of SEQ ID NO:2, the valine at residue 66 of SEQ ID NO:2, the glycine at residue 88 of SEQ ID NO:2, or the tryptophan at residue 117 of SEQ ID NO:2; and

and instructions for a diagnosing hearing disorder by amplifying all or a portion of SEQ ID NO:1 such that ~~all or a portion of exon 4 and exon 5 is amplified~~ one or more nucleotides encoding one or more of an amino acid at residue 51, an amino acid at residue 66, an amino acid at residue 88 and an amino acid at residue 117 of SEQ ID NO:2 is amplified and detecting ~~a lesion which is an insertion, a deletion, or~~ the absence or presence of a substitution of one or more nucleotides encoding one or more of a proline at residue 51 of SEQ ID NO:2, a valine at residue 66 of SEQ ID NO:2, a glycine at residue 88 of SEQ ID NO:2, and a tryptophan at residue 117 of SEQ ID NO:2.

44. (Previously presented) The kit of claim 43, wherein the hearing disorder is DFNA9.

45. (Previously presented) The kit of claim 43, wherein the probe is a labeled probe.

46. (Previously presented) The kit of claim 43, wherein the kit comprises two or more probes and at least one of the probes is a labeled probe.

47. (Previously presented) The kit of claim 43, wherein the probe is at least 12 nucleotides in length.

48. (Previously presented) The kit of claim 43, wherein the probe comprises at least 12 consecutive nucleotides of SEQ ID NO:1.

49. (Amended) A nucleic acid primer for diagnosing a hearing disorder which hybridizes under stringent conditions to a portion of the nucleic acid sequence ~~comprising~~ of SEQ ID NO: 1 or complement thereof, wherein the primer amplifies all or a portion of exons 4 and 5 of SEQ ID NO:1 such that one or more nucleotides encoding one or more of an amino acid at residue 51, an amino acid at residue 66, an amino acid at residue 88 and an amino acid at residue 117 of SEQ ID NO:2 is amplified.

50. (Previously presented) The nucleic acid primer of claim 49, wherein the primer amplifies a portion of exon 4 that comprises nucleic acids encoding a proline at residue 51 of SEQ ID NO:2.

51. (Previously presented) The nucleic acid primer of claim 49, wherein the primer amplifies a portion of exon 4 that comprises nucleic acids encoding a valine at residue 66 of SEQ ID NO:2.

52. (Previously presented) The nucleic acid primer of claim 49, wherein the primer amplifies a portion of exon 5 that comprises nucleic acids encoding a glycine at residue 88 of SEQ ID NO:2.

53. (Previously presented) The nucleic acid primer of claim 49, wherein the primer amplifies a portion of exon 5 that comprises nucleic acids encoding a tryptophan at residue 117 of SEQ ID NO:2.

54. (Previously presented) The nucleic acid primer of claim 49, wherein the hearing disorder is DNFA9.

55. (Previously presented) The nucleic acid primer of claim 49, wherein the primer is at least 12 nucleotides in length.

56. (Previously presented) The nucleic acid primer of claim 49, wherein the primer comprises at least 12 consecutive nucleotides of SEQ ID NO:1.

57. (Amended) A nucleic acid probe for diagnosing a hearing disorder which hybridizes under stringent conditions to the complement of SEQ ID NO:1 or ~~naturally occurring variants thereof comprising a lesion~~ a nucleic acid sequence that differs from the SEQ ID NO:1 ~~lesion~~ at one or more nucleotides encoding one or more of a proline at residue 51 of SEQ ID NO:2, a valine at residue 66 of SEQ ID NO:2, a glycine at residue 88 of SEQ ID NO:2, and a tryptophan at residue 117 of SEQ ID NO:2~~, wherein the probe hybridizes to a portion of the complement of SEQ ID NO:1, or the naturally occurring variant, such that a lesion at one or more nucleotides encoding: a proline at residue 51 of SEQ ID NO:2, a valine at residue 66 of SEQ ID NO:2, a glycine at residue 88 of SEQ ID NO:2, or a tryptophan at residue 117 of SEQ ID NO:2 is detected~~ and which detects the absence or presence of a substitution at one or more nucleic acids that encode the proline at residue 51 of SEQ ID NO:2, the valine at residue 66 of SEQ ID NO:2, the glycine at residue 88 of SEQ ID NO:2, or the tryptophan at residue 117 of SEQ ID NO:2.

58. (Amended) The nucleic acid probe of claim 57, wherein the probes hybridizes to a portion of the complement of SEQ ID NO:1 or ~~the naturally occurring variant~~ the nucleic acid such that a lesion at one or more nucleic acids encoding a proline at residue 51 of SEQ ID NO:2 is detected.

59. (Previously presented) The nucleic acid probe of claim 57, wherein the probe is labeled.

60. (Previously presented) The nucleic acid probe of claim 57, wherein the hearing disorder is DFNA9.

61. (Previously presented) The nucleic acid probe of claim 58, wherein the probe detects a lesion at nucleotide 207 of SEQ ID NO:1.

62. (Amended) The nucleic acid probe of claim 57, wherein the probes hybridizes to a portion of the complement of SEQ ID NO:1 or ~~the naturally occurring variant~~ the nucleic acid such that a lesion at one or more nucleic acids encoding a proline at residue 66 of SEQ ID NO:2 is detected.

63. (Previously presented) The nucleic acid probe of claim 62, wherein the probe detects a lesion at nucleotide 253 of SEQ ID NO:1.

64. (Amended) The nucleic acid probe of claim 57, wherein the probes hybridizes to a portion of the complement of SEQ ID NO:1 or the ~~naturally occurring variant~~ nucleic acid such that a lesion at one or more nucleic acids encoding a proline at residue 88 of SEQ ID NO:2 is detected.

65. (Previously presented) The nucleic acid probe of claim 64, wherein the probe detects a lesion at nucleotide 319 of SEQ ID NO:1.

66. (Amended) The nucleic acid probe of claim 57, wherein the probes hybridizes to a portion of the complement of SEQ ID NO:1 or the ~~naturally occurring~~

~~variant~~ nucleic acid such that a lesion at one or more nucleic acids encoding a proline at residue 117 of SEQ ID NO:2 is detected.

67. (Previously presented) The nucleic acid probe of claim 66, wherein the probe detects a lesion at nucleotide 405 of SEQ ID NO:1.

68. (Previously presented) The nucleic acid probe of claim 57, wherein the probe is at least 12 nucleotides in length.

69. (Previously presented) The nucleic acid probe of claim 57, wherein the probe comprises at least 12 consecutive nucleotides of SEQ ID NO:1.